

A 40-day-old Infant with Cyanosis and Lung Infiltration and Congestive Heart Failure⁺ — Case SNUCH CPC-36 —

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CASE PRESENTATION

This 40-day-old boy was transferred to Sowha Children's Hospital for further evaluation of dyspnea and pallor on September 17, 1990. He was an orphan who was brought to the Children's Welfare Fund (CWF) Hospital at the age of 7 days. No other prenatal or perinatal history was available. He was managed at the CWF Hospital for pneumonia, jaundice and congestive heart failure without improvement.

At the time of admission to Sowha Children's Hospital the patient weighed 2440 gm. Head circumference was 34 cm and chest circumference was 30 cm. Heart rate was 128 per minute, respiratory rate 60 per minute and body temperature 37.2°C. Blood pressure was 65/35 mmHg. He was generalized pale and weak. Slight but definite generalized cyanosis and poor skin turgor were noted. The anterior fontanel was open. The face was symmetric and showed no dysmorphic features. The neck was normal. Although lung expansion was satisfactory there was a prominent intercostal retraction. Rhonchi and rales were audible over the entire lung fields. Heart rate was regular. A grade 3/6 systolic murmur was heard along the left sternal border.

The abdomen was soft. The liver was palpated 3 cm below the right costal margin. The extremities and genitourinary system were unremarkable. A chest radiograph showed cardiomegaly, increased pulmonary vascularity, and right upper lobe atelectasis with pneumonia. The arterial blood gas analysis at admission showed pH 7.48, pCO₂ 49 mmHg, pO₂ 33 mmHg, HCO₃ 35.9 mmol/l, BE 12.9 mmol/l, O₂ saturation 66.6%. Blood electrolytes were Na 129 mmol/l, K 3.9 mmol/l and Cl 89 mmol/l. Peripheral blood cell counts were hemoglobin 12.7 g/dl, hematocrit 39%, WBC 16,000/cmm(seg 69%, lymph 31%) and platelet 236,000/cmm. Blood chemistries were AST 74 IU/l, ALT 32 IU/l, BUN 32 mg/dl, and Creatinine 1.2 mg/dl. Calcium, phosphorus, protein and albumin were all within normal limits. C-reactive protein was positive.

After admission he was managed with an oxygen hood, antibiotics and digitalization. On the second day in hospital he had a bout of high fever. A blood transfusion was also done for the anemia. A subsequent chest radiograph showed an improvement of the cardiomegaly and right upper lobe pneumonia. There was a new hilar infiltration. On the third day in hospital echocardiography was performed. It showed features of congenital heart disease. After that procedure, he was placed on diuretics. However, the dyspnea became worse, and the heart remained enlarged on repeated chest radiographs.

On the fourth day in hospital his general condition appeared stable without fever. However, early in the next morning his respiration became irregular. After a brief episode of clonic seizure he sustained a cardiorespiratory arrest. After a resuscitation

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was done his blood $p\text{CO}_2$ increased to 67 mmHg. He was put on a respirator with IPPV. Hemoglobin was 10.7 g/dl, hemarocrit 32%, WBC 19400/cmm (band 7%, seg 69%, lymph 23%), platelet 146,000 /cmm. Prothrombin time was 17.3 sec (control 11.3 sec) and partial thromboplastin time was 57.4 sec (control 26-40 sec). Blood culture grew coagulase negative staphylococcus, urine grew acinetobacter calcoaceticus var anitratus, and endotracheal aspirate grew pseudomonas maltophilia. The antibiotics were changed to Ceftazidim and amkin. BUN and creatinine were 70 mg/dl and 1.9 mg/dl, respectively. Digoxin level at this time was 3.4 ng/ml. The digoxin dosage was reduced. On the sixth day in hospital blood gas data became worse despite the assisted ventilation. Blood $p\text{CO}_2$ was persistently above the level of 70 mmHg. Fibrin degradation product was positive by 1 : 80 and fibrinogen was 106 mg/dl. AST and ALT levels rose to 438 IU/l and 526 IU/l, respectively. He developed focal seizure. Electroencephalography showed a periventricular hypoxic lesion. His general condition became gradually worse and he died on the seventh day in hospital.

DISCUSSION

Dr. Yoon: At the time of admission the patient was less than 3 percentiles both in weight and head circumference. Coupled with a deficient past history there were no physical stigmata to estimate the gestational period of this infant, such as plantar crease or figure of ears, etc. Therefore, I cannot assume if he is a premature or low birth weight infant. I guess that his main problems were pneumonia and cyanotic congenital heart disease based on the findings of tachypnea, intercostal retraction, rhonchi and rale, heart murmur, cyanosis, and a palpable liver. A chest radiograph showed cardiomegaly, increased pulmonary vascularity, and right upper lobe atelectasis with pneumonia. There is no indication whether the increased pulmonary vascularity was of arterial or venous origin. The arterial blood gas analysis revealed metabolic alkalosis due to the effort of respiratory compensation. Hyponatremia and hypochloremia may have been due to diuretics. Complete blood counts re-

vealed mild anemia and leukocytosis which were probably related to the pneumonia. There were mildly increased level of transaminase, and increased levels of BUN and creatinine indicating mild to moderate renal impairment. EKG was not available. However, echocardiography (Fig. 1) showed a normal situs and connection, secundum type of ASD, enlarged right atrium and right ventricle, small left atrium and left ventricle, and common pulmonary venous chamber beyond the left atrium on the subcostal view. The draining site of this anomalous vein is uncertain but the intracardiac type can be excluded.



Fig. 1. Echocardiograph of the patient.

He was managed with digitalis and diuretics for heart failure, and antibiotics for pneumonia. But his condition deteriorated, and seizure and cardiorespiratory arrest developed on the fifth day in hospital. Thereafter DIC and sepsis developed and renal impairment progressed. He died of multi-organ failure due to DIC.

I would like to discuss this case in the context of two viewpoints; one is of the cyanotic congenital heart disease associated with increased pulmonary vascularity, and second is of congestive heart failure which developed early in the neonatal period. Many structural anomalies can be considered in this patient based on clinical findings. They are a double outlet right ventricle without pulmonary stenosis, truncus arteriosus, transposition of great arteries with large ventricular septal defects, no

PS, hypoplastic left heart syndrome, pulmonary veno-occlusive disease and total anomalous pulmonary venous return (TAPVR). However, based on the echocardiographic findings, the first four anomalies are excluded because of the normal relationship of the great arteries and the presence of four chambers. Usually, patients with hypoplastic left heart syndrome succumb earlier than this patient, pulmonary veno-occlusive disease presents unilaterally and cyanosis is not definite. Therefore, I think that TAPVR is the most probable anomaly in this patient. Additionally, an obstruction may exist in the pulmonary venous draining system because this patient had developed cyanosis and severe pulmonary symptoms very early in his life.

Dr. Yoon's diagnosis:

1. Total anomalous pulmonary venous return with uncertain draining site, but intracardiac type is excluded.
2. Congestive heart failure
3. Pneumonia
4. Sepsis
5. Disseminated intravascular coagulation

Pathology findings:

Postmortem examination revealed major anomalies limited to the chest organs. The heart was enlarged. The brachiocephalic branches were normally distributed and the great arteries were in normal relationship. There was a large anomalous vessel connecting pulmonary veins to the right superior vena cava via the innominate vein. Four pulmonary veins formed a common pulmonary vein which was connected to the innominate vein (Fig. 2). At the point of confluence of the pulmonary veins there was a definite narrowing of the lumen indicating stenosis. The superior vena cava was large and so was the right atrium. The foramen ovale was open and the left atrium was hypoplastic. An ostium secundum type atrial septal defect was noted. The right ventricle was hypertrophic and the outflow tract was unremarkable. The pulmonary artery

showed a normal branching pattern and the ductus arteriosus was closed. The left ventricle was small but no ventricular septal defect was present.

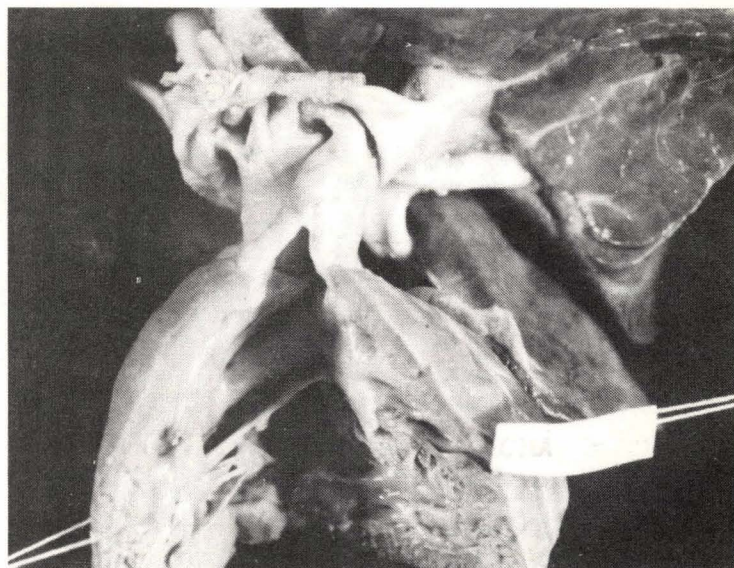


Fig. 2. Heart specimen shows an anomalous vein connecting pulmonary veins to the innominate vein to reach the right superior vena cava. The right ventricle shows hypertrophy.

The lungs were diffusely congested and the alveoli were filled with edema fluid. There was also a confluent bronchopneumonia involving both lungs. The liver was enlarged and showed acute passive congestion with centrilobular necrosis, suggestive of congestive heart failure. The kidneys showed severe acute tubular necrosis. The brain showed hypoxic ischemic encephalopathy with findings of periventricular leukomalacia. Postmortem blood culture grew coagulase negative staphylococci.

Final pathological diagnosis:

1. Total anomalous pulmonary venous return, supracardiac type, common pulmonary vein connecting to the innominate vein.
2. Confluent bronchopneumonia
3. Acute tubular necrosis
4. Sepsis (coagulase negative staphylococci)
5. Hypoxic-ischemic encephalopathy with periventricular leukomalacia